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Case Report

Coarctation of the thoracic aorta masquerading as bilateral aorto-iliac stenosis

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Summary We present a case with coarctation of the aorta (CoA) with lifestyle limiting claudication and lower extremity weakness, successfully treated with surgical correction. The presented case discusses the diagnostic challenges associated with identifying CoA in patients with claudication.

Introduction

Coarctation of the aorta (CoA) refers to a narrowing of the aortic lumen which can be located in the descending thoracic aorta, the middle aorta, or even the infrarenal aorta. CoA of the descending thoracic aorta is most commonly classified as a congenital cardiovascular anomaly which accounts for 7% of congenital heart disease [1]. Patients with unrepaired CoA may present with a variety of chronic manifestations including upper extremity hypertension, ventricular hypertrophy, and eventual cardiac failure [2—6]. Furthermore, reduced blood flow through the constricted aorta may manifest as lower extremity claudication or reduced renal perfusion. We present a case of de novo diagnosis of CoA in an adult presenting with symptoms of lifestyle limiting claudication due to decreased lower extremity perfusion. The literature regarding occurrence of claudication in this context is reviewed.

Case report

A 41-year-old Hispanic non-smoker male with a significant past medical history of coronary artery disease, history of myocardial infarction followed by percutaneous coronary intervention and ischemic cardiomyopathy had an ejection fraction (EF) of 15%. The patient's prior medical care was fairly non-existent and he established his medical care in our institute for his heart failure. During the regular follow-ups, the patient's medical therapy and neurohormonal blockade for his heart failure was optimized. Nevertheless, he complained of progressive bilateral exertional buttock and thigh claudication. Peripheral pulse exam demonstrated diminished common femoral, popliteal, and dorsalis pedis pulses bilaterally. The patient underwent a diagnostic workup, including lower extremity arterial segmental physiologic

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Table 1 Segmental arterial pressure of the lower extremity. Pre- and post-surgical repair of the coarctation of the aorta.

Pre-surgical repair LE segmental blood pressure					Post-surgical repair LE segmental blood pressure				
Extremity	Right		Left		Extremity	Right		Left	
	BP	LE/UE ratio	BP	LE/UE ratio		BP	LE/UE ratio	BP	LE/UE ratio
Arm	127		126		Arm	95		86	
Upper thigh	103	0.81	117	0.92	Upper thigh	82	0.86	87	0.92
Lower thigh	89	0.70	104	0.82	Lower thigh	100	1.05	95	1.00
Calf	88	0.69	112	0.88	Calf	87	0.92	87	0.92
Ankle	84	0.66	83	0.65	Ankle	86	0.91	90	0.95

BP, blood pressure; LE, lower extremity; UE, upper extremity.

evaluation with ankle-brachial index (ABI) (Table 1). The patient's history and segmental pulse exam were suggestive of significant bilateral aorto-iliac arterial stenoses. Given the above findings, the patient was referred for lower extremity peripheral angiography.

The peripheral angiogram demonstrated no obstructive lower extremity peripheral arterial disease in either leg. Relatively sluggish three-vessel run-off to the foot was present bilaterally. Notably, the arterial waveform was dampened in the distal abdominal aorta when compared to the central ascending aortic pressure. To confirm the presence and evaluate the level of the gradient, a gradual pullback was performed from the ascending aorta to the descending aorta using a 5-French pigtail catheter (Fig. 1). There appeared to be a 25 mm Hg gradient across the descending aorta just distal to the left subclavian artery. An aortic arch aortogram, with the pigtail catheter placed in the ascending aorta, was performed using a power injection of contrast and confirmed the presence of CoA (Fig. 2). In retrospect, the patient's chest X-rays were reviewed and they did not show any of the classic findings of CoA.

After discussion with the patient regarding endovascular treatment versus open repair, the patient elected to undergo surgical correction. Impacting this decision was the need for concomitant surgical pacemaker lead extraction. The patient underwent a 16-mm endograft placement with end-to-side anastomosis from the proximal to the distal segment of the coarctation bypassing the stenosed segment on partial cardiopulmonary bypass. Post operatively the patient had complete resolution of his claudication symptoms and normalization of his ankle brachial indices and segmental pressures (Table 1). The interval transthoracic echocardiography after the surgical correction showed no significant change in ventricular function with an EF of 12% and a severely dilated left ventricle.

Discussion

CoA is most commonly a congenitally acquired anatomic obstruction. CoA arises from abnormal development of the embryonic left fourth and sixth aortic arches and usually manifests during childhood and early adulthood, although it can remain asymptomatic for prolonged periods [7—9]. In patients with unrepaired CoA, life expectancy is reduced due to the development of a systemic hypertension equivalent state and its associated complications. The diagnosis of CoA may be missed in children and contribute to significant morbidity during adulthood. The diagnosis is often discovered when a work up of secondary hypertension is begun, or as an incidental finding on chest imaging. In part, the high prevalence of systemic hypertension in the population and the infrequent routine measurement of lower extremity blood pressures make this a challenging disorder to identify. Classic findings on chest X-ray such as a widened cardiac silhouette, wide ascending and descending aorta, and notching of the ribs may not always be present or evident as in our patient [10,11]. Comprehensive examination using upper and lower extremity blood pressures, echocardiography, computed tomography angiography (CT), or magnetic resonance imaging (MRI) is paramount to identifying the nature of the CoA. In the present case invasive hemodynamics during catheterization followed by digital subtraction angiography was invaluable at identifying the presence and severity of the CoA.

Variants of thoracic CoA have been described in the context of claudication. These include congenital distal or mid-aortic occlusions and acquired CoA from thrombosis and atherosclerosis. Distal aortic or middle aortic syndromes can present in infants and children as hypertension, renal insufficiency, and lower extremity atrophy or claudication [12,13]. Middle aortic syndrome rarely presents as a de novo finding in adults and the majority of cases are under the age of 30 [14]. These lesions are generally attributed to congenital hypoplasia. In adults, an atherosclerotic etiology of middle or distal aortic stenosis should be considered. For example, Chen et al. described a case of discrete distal aortic stenosis in a 63-year-old female smoker with claudication who underwent surgical repair [15]. The pathologic analysis of the stenotic region demonstrated extensive cholesterol deposits with proliferation of fibroblasts and smooth muscles — all consistent with severe atherosclerosis. In its most aggressive form, aortic atherosclerosis may result in progressive narrowing of the aortic lumen leading to total occlusion [16]. This process is often accompanied by thrombosis and the development of organized thrombus. Chin et al. described a case of a 69-year-old male smoker with a history of claudication and resistant hypertension who presented with a functional coarctation of the aorta secondary to extensive thrombus extending into the aorto-iliac bifurcation [17]. Sheikhzadeh et al. reported two cases of acquired thromboatheromatous CoA which presented with claudication in

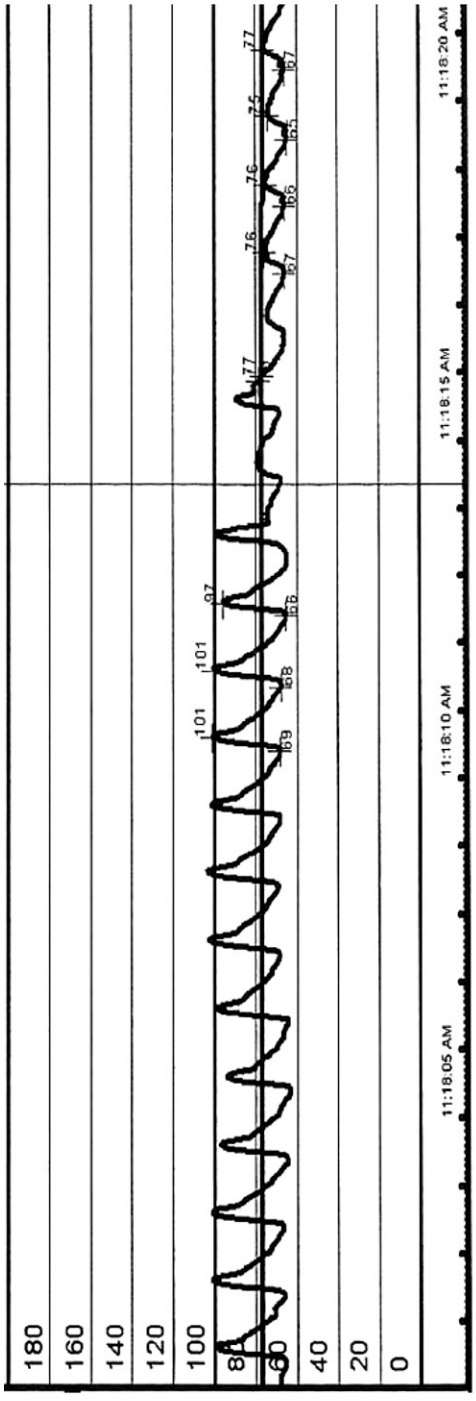

Figure 1 Pullback gradient with a 5-French pigtail catheter across the coarctation demonstrating a 25 mm Hg pressure gradient.

Figure 2 Digital subtraction image of the coarctation of the aorta. White arrow points to region of coarctation of the aorta.

patients with a smoking history [18]. In both cases, visible (on CT scan) thrombus was present in the descending thoracic aorta resulting in obstruction at or just distal to the left subclavian. In these cases, the clinical presentation, CoA location, and intravascular pressure measurements were very similar to congenital thoracic CoA. Distinguishing these two rare phenomenon are obviously important as the treatment plan will markedly differ in regards to endovascular or surgical repair, anticoagulation, antiplatelets, and lipid-lowering therapies.

Treatment of CoA is increasingly focused on endovascular angioplasty and stenting. Recent data would suggest that both intermediate and long-term outcomes of patients following stenting are comparable to surgically treated individuals [19]. A given patient's age, coarctation morphology/location, coexistent co-morbidites, and prior history of aortic interventions should all be considered while determining the optimal treatment option for each patient. Options for surgical correction include resection of the affected aortic segment and end-to-end anastomosis, patch aortoplasty, left subclavian flap angioplasty, and bypass grafting [20—22]. Resection of the coarctation site and end-to-end anastomosis is a preferred surgical method since it preserves the left subclavian artery, which is important in adolescents and adults, and avoids usage of prosthetic materials. Patch aortoplasty has become less popular and is rarely used due to higher incidence of aneurysmal dilatation [23]. Bypass graft is also utilized and some institutions use left subclavian-aortic bypass grafting as the primary approach in all adult coarctation cases owing to its simplicity and low risk [22].

The present case demonstrates the importance of considering CoA in the differential diagnosis for claudication. Segmental pressures may suggest aorto-iliac disease in these cases. Careful attention to invasive hemodynamics with low

threshold to obtain further imaging must be considered in such cases.

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